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Factors influencing Pseudomonas persistence in intermittently infected cystic fibrosis (CF)-patients

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Aims: Determination of factors influencing the persistence of Pseudomonas aeruginosa in sputum or oropharyngeal cultures in CF-patients who are not chronically infected.
Methods: 187 CF-patients, mean age 16.7, were classified with regard to their status of Pseudomonas infection (chronic, intermittent, not infected). 24 patients with 1-7 positive cultures during a 2 year period were identified with intermittent infection. Appropriate anti-Pseudomonas therapy was administered as clinically required. Duration of intermittent infection, frequency of Pseudomonas detection, and number of different pyocin typed strains were analysed retrospectively. The cohort was prospectively observed with three monthly cultures over the next two years.
Results: In 9 patients Pseudomonas was not detected within the next two years, 15 patients showed unchanged intermittent infection. The disappearance of Pseudomonas depended on the total duration of intermittent Pseudomonas infection (3.6 vs 6.6 years, p=0.06), the frequency of positive Pseudomonas cultures (3.8 vs 11.9, p=0.002), the number of different strains (2.0 vs 4.4, p=0.03), and the absence of antibodies against Pseudomonas (7/9 vs 5/15, p=0.036). If Pseudomonas disappeared, FEV-1 was stable in the following two years, if not, FEV-1 dropped (98 to 88.5 predicted, p=0.028). Anti Pseudomonas therapy was less intense during the two prospective years in patients where Pseudomonas disappeared (2/9 vs 9/15 cycles of iv therapy, p=0.021).
Conclusions: Disappearance of Pseudomonas aeruginosa from intermittently infected CF-patients is more likely with shorter duration of infection, less positive cultures, less different Pseudomonas strains and the absence of antibodies against Pseudomonas. Patients with ongoing intermittent infection required more intense anti-Pseudomonas therapy and nevertheless had deteriorating lung function.

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Factors associated with increased rate of CF pulmonary exacerbations in the South and West of England

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Aims: Pulmonary exacerbations (P Exs) are important feature of CF. They are expensive, and associated with poor quality of life. This study examines factors associated with P Exs in a large cohort in adolescent and adult patients.
Patients: This is a cross-sectional study on data collected in the South and West Regions in England in 2002. Patients over 12 years were include. Data on age, gender, BMI, genetic profile, Swat test results, FEV1, infection with Pseudomonas aeruginosa (Pa) and on CF-related diabetes were included. Exacerbation was regarded to be present when IV antibiotics were provided. Patients were divided into 4 groups: 1 (223 patients), no exacerbations; group 2 (158 patients); 1-2 annual exacerbations; group 3 (77 patients), 3-4 annual exacerbations; and group 4 (49 patients) more than 4 exacerbations in a year.
Results: The need for IV antibiotics increased with decline in FEV_1, the presence of Pa in the sputum, CF-related diabetes and with body mass index below 20. Rate of pulmonary exacerbations did not differ according to patients' age or gender. There was no difference in either the concentration of sweat sodium or chloride concentration or genetic profile across categories of exacerbations.
Conclusions: Reduced FEV_1, infections with Pa, reduced BMI and CF related diabetes are associated with increased pulmonary exacerbations as identified by the need of greater number of annual courses of IV antibiotics.

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The use of mobile technologies in the early detection of pulmonary exacerbation of CF. Time to be precise!

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Aims: Pulmonary exacerbation is an important outcome in CF. Defining exacerbations differ between physicians. The aim of this study is to investigate change in symptoms and FEV1 in patients in the period around CF pulmonary exacerbation.
Methods: We use device consisting of a portable computer/mobile phone attached to a spirometry. Patients are asked to complete once daily a symptom questionnaire scoring their symptoms of cough, sputum, breathlessness and fatigue and to perform spirometry on 3 occasions. Data is sent on a daily basis and sought by a group of researchers. Data produced on the first exacerbation-free 20 days are regarded as baseline data. Exacerbations were regarded to have occurred if patient sought help from the CF team, or if they have 3 days increase in three respiratory symptoms, or two respiratory symptoms and 10% decline in FEV1 or three days of decline in FEV1 alone.
Results: So far, 19 patients were enrolled in the study. Their mean age 24.2 years, range 19-34 years. Mean days recorded 67.68 % of total. From a total of 22 exacerbations that were recorded, 14 patients registered an increase in one symptom and for 13 patients there was an increase of 2 symptoms. A decline in FEV1 occurred in 4 patients. The prodromal phase varied between patients. In 22 exacerbations recorded by the e-san system, a course of antibiotics was required.
Conclusions: Our criteria for exacerbation in this cohort of adult CF patient may have been too stringent. The need for treatment as felt by patient can occur by increase in one or two symptoms. Precise definition of CF pulmonary exacerbation will need more data from this.

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Identification of factors affecting the survival in patients infected with Burkholderia cenocepacia

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Up to 25 % of patients attending the Prague CF Centre are infected with B. cenocepacia (BC; formerly genomovar III) of the Burkholderia cepacia complex. Despite this genomovar and strain homogeneity, the infection is manifested in its whole spectrum from its slow progression to the fulminant cepacia syndrome. Therefore, we sought to define the clinical factors influencing the survival after B. cenocepacia infection.
Within 1994 - 2004, in total 96 patients were infected with BC. Using standard statistical tests (Cox model, Gehan-Wilcoxon, Cox-Mantel and Ln ranks tests), we analysed the following factors in terms of their possible role in patients survival: lung function at the time of BC infection, change of FEV_1 per year during the first three years, Pseudomonas aeruginosa infection, nutritional status, pancreatic status, sweat chloride concentration, CFTR genotype and gender.
We found statistically significant affect of the lung function (FEV_1) at BC infection ($p<0.001$, $b=-0.057$, i.e. $e^b=0.947$), of dynamic changes in FEV_1 ($p<0.001$, $e^b=0.741$), and of chloride concentration ($p=0.03$, $e^b=1.025$) on the survival. Influence of nutritional status, previous P. aeruginosa infection and gender was not proven. The role of CFTR genotype and pancreatic status were not reliably evaluated due to a small number of non-censored observations in the group with mild mutations and pancreatic sufficiency (p value was close to 0.05).
Results are in an agreement with published data about bad lung functions and their further decline worsening the infection prognosis. Our data indicate that patients with milder impairment of CFTR function may have less severe course of the BC infection.
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